



UNIVERSITÀ
DEGLI STUDI
FIRENZE

FLORE

Repository istituzionale dell'Università degli Studi di Firenze

Imaging studies in extramedullary hematopoiesis of the spleen

Questa è la Versione finale referata (Post print/Accepted manuscript) della seguente pubblicazione:

Original Citation:

Imaging studies in extramedullary hematopoiesis of the spleen / B. Matteuzzi; L. Pieri; F. Lisi; S. Galimberti; D. Campani; S. Colagrande. - In: ANNALS OF HEMATOLOGY. - ISSN 0939-5555. - ELETTRONICO. - (2013), pp. 1-3. [10.1007/s00277-013-1802-5]

Availability:

This version is available at: 2158/814962 since:

Published version:

DOI: 10.1007/s00277-013-1802-5

Terms of use:

Open Access

La pubblicazione è resa disponibile sotto le norme e i termini della licenza di deposito, secondo quanto stabilito dalla Policy per l'accesso aperto dell'Università degli Studi di Firenze (<https://www.sba.unifi.it/upload/policy-oa-2016-1.pdf>)

Publisher copyright claim:

(Article begins on next page)

Imaging studies in extramedullary hematopoiesis of the spleen

Benedetta Matteuzzi • Lisa Pieri • Francesca Lisi •
Sara Galimberti • Daniela Campani • Stefano Colagrande

Received: 22 April 2013 / Accepted: 18 May 2013
© Springer-Verlag Berlin Heidelberg 2013

Dear Editor,

A 63-year-old man underwent CT studies before surgery for an aortic aneurysm. Contrast medium-enhanced CT scan showed an increased spleen size with a solid, 10 cm large, round, well-defined highly enhanced lesion, suggesting high vascularity with nonhomogeneous density due to the presence of some pseudocystic areas (Fig. 1a) [1]. An increased spleen size had been discovered 13 years before without any symptoms and/or abnormality in complete blood counts (CBCs). Three years later, MRI studies confirmed a slightly increased lesion (12 cm) with a thin hypointense halo and a heterogeneous hyperintense signal intensity on T2-weighted images (i.e., well hydrated) (Fig. 1b, c). Diffusion-weighted

images showed patterns compatible with a high cellularity (Fig. 1d). So, malignancy could not be excluded, and the patient underwent splenectomy.

Histological examination was consistent with extramedullary hematopoiesis, characterized by sinusoidal dilatation and hypolobated megakaryocytes often gathered in clusters (Fig. 1e, f). CBC and blood smears remained within the normal range. However, bone marrow examination showed an increased cellularity of 70 % with trilineage hyperplasia, with no increased fibrosis or blasts. The search of JAK2 V617F mutation was negative. Even if clinical and hematological findings did not fulfill the WHO 2008 diagnostic criteria for any of the myeloproliferative neoplasms [2], histological pattern of the bone marrow was suggestive for polycythemia vera, and hematopoiesis in the spleen had features resembling myelofibrosis. Two years after splenectomy, evidence of a myeloproliferative neoplasm did not occur.

Extramedullary hematopoiesis can be a compensatory response to hyperproliferating bone marrow cells, as is the case of hemoglobinopathies such as thalassemia syndromes, sickle-cell anemia, and other chronic hemolytic anemias, or can occur in patients with chronic myeloproliferative neoplasms, especially myelofibrosis. Foci of extramedullary hematopoiesis can be found in various organs, even if it predominantly affects the liver and spleen. The localizations of extramedullary hematopoiesis may be also in the thoracic paravertebral areas, lymph nodes, adrenal gland, retroperitoneal fat, and renal pelvis [3, 4].

To our knowledge, only a few cases of focal extramedullary hematopoiesis have been reported with imaging descriptions [5]. In fact, spleen tumors are usually detectable but not easily characterizable by imaging due to overlapping patterns between benign and malignant nodules. The greatest part of neoplasms of the spleen is benign: hemangiomas, inflammatory

B. Matteuzzi • F. Lisi • S. Colagrande
Department of Experimental and Clinical Biomedical Sciences,
Radiodiagnostic Unit n. 2, Azienda Ospedaliero-Universitaria
Careggi, University of Florence, Florence, Italy

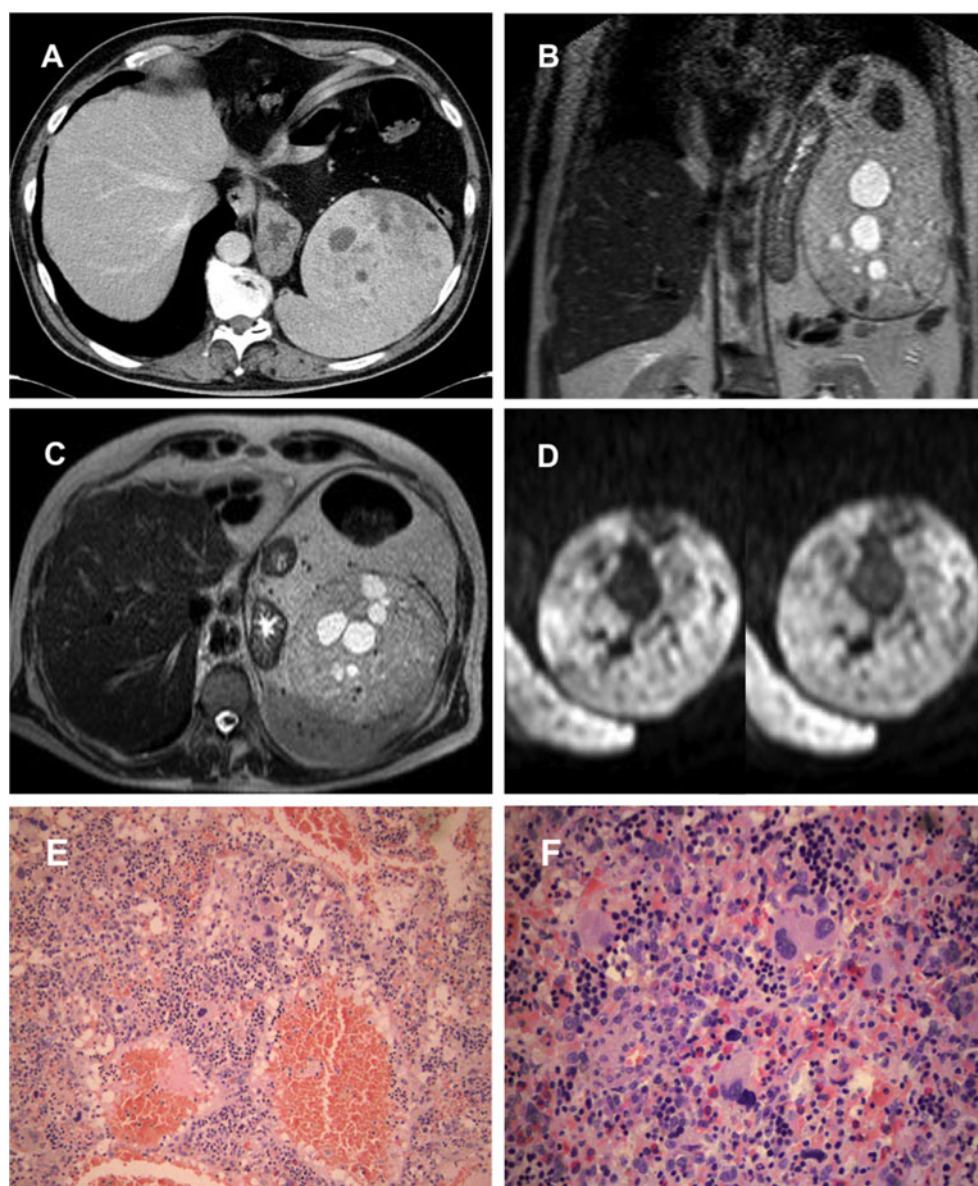
L. Pieri
Department of Experimental and Clinical Medicine, Section
of Haematology, Azienda Ospedaliero-Universitaria Careggi,
University of Florence, Florence, Italy

S. Galimberti
Department of Experimental and Clinical Medicine, Section
of Haematology, Azienda Ospedaliero-Universitaria di Pisa,
University of Pisa, Pisa, Italy

D. Campani
Department of Surgical Pathology, Molecular, Medical and Critical
Care, Azienda Ospedaliero-Universitaria di Pisa, University of
Pisa, Pisa, Italy

S. Colagrande (✉)
Department of Biomedical Sciences and Clinics, Radiology
Unit n. 2, Azienda Ospedaliero-Universitaria Careggi, University
of Florence, Largo Brambilla 3, Florence, Italy 50134
e-mail: stefano.colagrande@unifi.it

Fig. 1 **a** CT scan after administration of intravenous contrast medium shows a large focal lesion with well-defined margin and heterogeneous density due to internal colliquative areas. **b, c** MRI, T2-weighted coronal (**b**), and axial (**c**) scans confirm the lesion with heterogeneous signal intensity, slightly hyperintense on T2-weighted images, with a thin hypointense halo. **d** MRI and heavily diffusion-weighted axial scans show high signal intensity and then a pattern suggestive for crammed cells. **e, f** Histological examination of the nodule, showing extramedullary hematopoiesis with ectasic sinusoids and clustered hypolobated megakaryocytes



pseudotumors, and splenic cysts are the most frequent [6, 7], while lymphoma is the most common of malignancies, followed by other less frequent histologic types as hemangioendotheliomas, hemangiosarcomas, and metastases [8]. Our report emphasizes the role of radiologic features for the diagnosis of intrasplenic extramedullary hemopoiesis, bearing in mind that splenic biopsy is a very risky procedure.

Even if none of the above-described findings is neither sufficient nor necessary for definitive diagnosis, the association of well-defined lesion, highly and inhomogeneously enhancing, with pseudocystic areas, and medium-high signal intensity in MRI—T2 and diffusion weighted—not resembling hemangioma or pseudotumor, could suggest radiologists a diagnosis

of intrasplenic extramedullary hemopoiesis [4, 5] and then clinicians to perform tests for hematological disorders including myeloproliferative neoplasms, such as bone marrow biopsy, JAK2V617F or MPL mutations in addition to CBC, before referring the patient for splenectomy which can bring some risks of complications.

Acknowledgments The study was in part funded by a grant from the Associazione Italiana per la Ricerca sul Cancro (AIRC, Milano) “Special Program Molecular Clinical Oncology 5×1000” to the AIRC-Gruppo Italiano Malattie Mieloproliferative (AGIMM), project number 1005. A detailed description of the AGIMM project is available at <http://www.progettoagimm.it>.

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Priola AM, Gned D, Boccuzzi F, Priola SM (2012) Unusual focal intrahepatic extramedullary haematopoiesis in alpha-thalassaemia. *Liver Int* 32:771
2. Tefferi A, Vardiman JW (2008) Classification and diagnosis of myeloproliferative neoplasms: the 2008 World Health Organization criteria and point-of-care diagnostic algorithms. *Leukemia* 22:14–22
3. Singer A, Maldjian P, Simmons Z (2004) Extramedullary hemopoiesis presenting as a focal splenic mass: case report. *Abdom Imaging* 29:710–712
4. Bradely MJ, Metreweli C (1990) Ultrasound appearance of extramedullary in the liver and spleen. *Br J Radiol* 63:816–818
5. Sohawon D, Lau KK, Lau T, Bowden DK (2012) Extra-medullary haematopoiesis: a pictorial review of its typical and atypical locations. *J Med Imaging Radiat Oncol* 56:538–544
6. Giovagnoni A, Giorgi C, Goteri G (2005) Tumor of the spleen. *Cancer Imaging* 5:73–77
7. Maternini M, Misani M, Vanzati A (2012) Extramedullary hemopoiesis and littoral cell angioma of the spleen: our experience and review. *Hepatogastroenterology* 59:1789–1793
8. Böcker W, Denk H, Heitz (2001) *Pathologie*, 2nd edn. Urban & Fischer, München, pp 537–538